260 Supra-maximal verification of peak oxygen uptake in adolescents with cystic fibrosis

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Aim: Several studies have reported a reduced peak oxygen uptake (VO_{2peak}) during incremental exercise testing in patients with cystic fibrosis (CF). The objective of this investigation was to verify the VO_{2peak} attained in traditional cardiopulmonary exercise testing (CPET) in adolescents with CF by using a supra-maximal exercise test.
Methods: Sixteen adolescents with CF (8 boys, 8 girls; mean age 14.6±1.7 yr; FEV_1%predicted [range 45–117%]) volunteered in this study and successively performed CPET and a supra-maximal test [Steep Ramp Test protocol (SRT)], using the same equipment, on the same day.
Results: CPET and the SRT resulted in comparable minute ventilation, breathing reserve and heart rate values. One-way repeated measures ANOVA revealed no significant difference in VO_{2peak}/kg between CPET and SRT (38.9±7.4 and 38.8±8.5 $mL \cdot min^{-1} \cdot kg^{-1}$, respectively; $p = 0.81$), and good to excellent associations were found for peak exercise parameters between both tests ([r 0.71–0.98]; $p < 0.01$). Peak work rate was significantly higher (~50%) in the SRT compared with CPET ($p < 0.01$). Bland–Altman plots show no systemic bias for CPET and SRT measurements of VO_{2peak}/kg. No differences were found between CPET and SRT VO_{2peak} values within and between the maximal and non-maximal effort group ($p > 0.4$) based on the Rowland criteria.
Conclusion: The VO_{2peak} measured in CPET seems to reflect the true VO_{2peak} in adolescents with CF.

261 Social benefits and peak work capacity in an adult CF population

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The complex UK benefits system, based on the degree of disability caused by disease, is increasingly being scrutinised and this has adversely affected some CF patients. Physical activity is important in CF, offsetting disease limitations and optimizing quality of life and thereby reducing disability: accurate measurement of this may ensure that patients receive the correct level of benefit. As a marker of this, we measured Peak Work Capacity (PWC%) (corrected for lung function) using incremental cycle ergometry in 35 CF adults (17 male) and compared it to the level of benefits received and an activity score calculated from a daily activity questionnaire.
Ten (29%) led a sedentary lifestyle and 21 (60%) a low active lifestyle, where the most common form of exercise was walking. Of the 34 who had exercise testing, 3 (9%) had marked and 11 (32%) moderate impairment of capacity. Severity of disease (FEV_1%) correlated strongly with PWC% ($r = 0.71$, $p = 0.03$), but correlation of benefits with lifestyle ($r = 0.18$, $p = 0.02$) and peak work capacity ($r = -0.41$, $p = 0.02$) were less marked (see table). This study shows that PWC% can be used to aid the evaluation of the effect of disease on everyday life, and hence the disability caused. Such a test may aid patients to obtain the correct level of benefit, thereby avoiding needless and distressing government scrutiny.

Table 1

Benefit	Males			Females		
	N	Mean FEV1 %	PWC %	N	Mean FEV1 %	PWC %
Low	2	66	60	1	109	120
Middle	4	73	71	4	72	70
High	11	60	62	13	52	60

262 Health related quality of life in adults with cystic fibrosis: the impact of exercise tolerance

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Health-related Quality of Life (HRQoL) plays a key role in the clinical course of cystic fibrosis (CF). However, there is a lack of data regarding the impact of physical fitness on HRQoL in CF patients. The present study examined the relationship between clinical variables, exercise capacity and domains of HRQoL. 54 adults with CF (FEV_1: 53±13%, age: 26±7 years, BMI: 21±2 kg/m^2) performed a six-minute walk test (6MWT) and completed the CF Questionnaire for adults (CFQ 14+). The CFQ 14+ includes three modules for assessing QoL, symptoms and health perception. The main outcomes were: walk distance % predicted (6MWD%), oxygen desaturation, heart rate reserve, FEV_1%, FVC% and BMI. The relationships between these parameters and domains of HRQoL were evaluated using Pearson's correlation and stepwise multiple-regression.
The 6MWD% was significantly correlated with 8 domains of HRQoL (Pearson's r ranged from 0.26 to 0.58). FEV_1% was significantly correlated with 4 domains of HRQoL (Pearson's r ranged from 0.26 to 0.43). Multiple-regression analysis revealed that the 6MWD% was the most significant predictor of the majority of HRQoL dimensions. It was notably independently responsible for 35% of the variance in respiratory symptoms and for 26% in health perception. The 6MWD% was a better predictor of HRQoL than the usual primary outcomes of disease severity. These results emphasize the clinical interest of the 6MWT. Exercise rehabilitation strategies designed to enhance submaximal exercise capacity have the potential to impact positively on different domains of HRQoL in CF patients.
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263* Breathlessness during exercise in cystic fibrosis

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Introduction: Breathlessness limits exercise in Cystic Fibrosis (CF). Complex disturbances in respiratory mechanics during exercise make it difficult to determine which alterations contribute most strongly to the sensation of breathlessness. This study aimed to examine interactions between breathlessness, metabolic and ventilatory variables, intrathoracic and intraabdominal pressures and neural respiratory drive (NRD), assessed using diaphragm EMG (EMGdi) during exercise.
Methods: Fifteen patients [mean(SEM) age 24(2) years] with CF [FEV_1 % predicted 53.5(6.2)%] performed incremental cycle exercise to exhaustion. During exercise respiratory flow, volume, metabolic data, oesophageal and gastric pressures were recorded and work of breathing was determined by measurement of the diaphragm pressure time product (PTPdi). EMGdi was recorded using a multipair oesphageal catheter. Inspiratory capacity (IC) and Borg scores were measured every minute.
Results: Mean(SEM) exercise duration was 20.5(1.2) minutes, maximal work rate achieved was 142(13.4) watt and peak VO_2/kg 28(2.2) ml/min. Patients became dynamically hyperinflated during exercise as indicated by a reduction in IC from 1.86(0.52) L at rest to 1.39(0.39) L at end exercise, $p = 0.002$. EMGdi amplitude increased progressively during exercise, whereas PTPdi and minute ventilation plateaued at 70% of the total exercise time, indicating neuromechanical dissociation. The intensity of breathlessness increased with the onset of neuromechanical dissociation, with Borg scores being more strongly correlated with EMGdi ($r = 0.988$, $p = 0.001$).
Conclusion: NRD was best correlated to breathlessness following the onset of neuromechanical dissociation.